Title: Ruptured pseudoaneurysm as a cause of spontaneous intracerebral bleed in a 3-month old infant

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Abstract

Ruptured intracranial aneurysms in infants are very rare but if missed can lead to poor outcomes. Spontaneously dissecting false aneurysms have been described only in a handful of cases. We report a case of a three-month old girl with deteriorating neurological function due to a ruptured distal middle cerebral artery pseudoaneurysm.

Introduction

Ruptured intracranial aneurysms are considered the second leading cause of intracranial hemorrhage (ICH) after arteriovenous malformations in the paediatric age group [1]. In infants' (< 1yr old), intracranial aneurysms also present with subarachnoid hemorrhage, seizures and direct mass effect (giant aneurysms) causing focal neurological deficit [2]. Intracranial aneurysms in this age group may however form a distinct subgroup with an incidence of less than 0.1% and a slight predilection for males (1.1:1 M:F ratio) [2]. The average age at presentation is 4.6 months with younger infants being more likely to present with haemorrhage [2]. The middle cerebral artery (MCA) is three times more commonly affected than any other cerebral vessel followed by anterior cerebral artery (ACA), internal carotid artery (ICA) and basilar artery (BA) [2]. Since only a small number of cases have been reported in the literature, the clinical presentation, pathogenesis and management is variable. We, herein, present a case of a surgically clipped MCA dissecting pseudo-aneurysm in a 3-month old girl. Her clinical presentation, management, follow up and recovery are discussed in the context of the available literature.

Clinical Presentation

A full term, three month old previously healthy girl was transferred to the paediatric neurosurgery service with a sudden onset of irritability, poor feeding, vomiting and generalised loss of tone. There was no history of trauma. Subsequently she developed seizures which settled on phenobarbitone. On arrival to the paediatric neurosurgical unit, her GCS was recorded as 13 out of 15. There was no pupillary asymmetry. The anterior fontanel was bulging. While there was no focal neurological deficit, she had generalised brisk reflexes.

A computerised tomography (CT) scan of the brain demonstrated an acute left frontal intracerebral bleed extending into the Sylvian fissure, causing mass effect. A magnetic resonance (MRI) scan of the brain showed a vascular abnormality in relation to the clot. A subsequent CT angiogram (CTA) of the brain demonstrated a 7mm pseudo-aneurysm of the left anterior M3 branch; the appearances were in keeping with arterial dissection. Postoperative vascular follow up imaging was deemed unnecessary in this particular case. The coagulation profile and platelet count were normal as was the ultrasound of the kidneys.

Endovascular intervention was deemed high risk. She therefore underwent surgical evacuation of the clot and the pseudo-aneurysm was excised by sacrificing the parent vessel with a proximal clip. She made a swift recovery and had no neurological deficit on discharge and at six months follow-up. A postoperative CT scan of her brain showed complete resolution of the clot with no infarcts. Histology confirmed features consistent with pseudo-aneurysm.

Discussion

Intracranial aneurysms have been reported in as little as 150 infants between 1943 and 2005 (see [2] for a review); Spontaneous non-traumatic and non-infectious aneurysms were seen in 77 cases only. In a more recent case-series study, one out their 74-paediatric intracranial aneurysms was reported in an infant (age=42 days) [3]. Taken their rarity, the characterization, clinical outcomes and length of follow up of such spontaneous intracranial aneurysms in this age group is yet to be fully determined.

The adult recognised risk factors for intracranial aneurysms such as smoking and hypertension are unlikely to contribute to the aetiology of aneurysms in infants. Like adults and older children, infectious and traumatic causes were however reported in approximately 10% and 15% of infant aneurysmal cases, respectively [2]. In the majority of cases, the exact aetiology remains unknown, however, the absence of lamina elastica interna, muscularis media or both was commonly reported.

Infant aneurysms range in size between 0.1-9cm with an average size of 1.8cm [2]. Smaller aneurysms (less than 2.5cm) are more often associated with haemorrhage at initial presentation; an observation that may support the hypothesis that intramural factors play a key role in the pathogenesis of infants aneurysms [2]. In our case report, the aneurysm size was only 7mm and therefore unsurprisingly presented with an intracranial haemorrhage in the absence of trauma.

Depending on the type, location, anatomical configuration, clinical presentation and availability of facilities and expertise, a wide range of

approaches have been used to treat infant aneurysms. These include expectant management with or without antibiotics (the later reserved for mycotic aneurysms), surgical clipping, endovascular embolization and ligation of the parent vessel. In this case report the aneurysm was distal enough to be completely excised whilst the parent vessel was clipped with no postoperative complications.

As in adults, a non-contrasted CT scan is the initial investigation when intracranial bleed is suspected in infants. In our case, the location of the haematoma and distribution of blood on the non-contrasted CT has prompted us to proceed with an MRI followed by a CTA to identify the source of the bleed. Based on the information retrieved from the MRI and CTA, formal catheter cerebral angiography was deemed unnecessary particularly with the radiation exposure risk at this age. The decision to operate was made after a multidisciplinary team discussion involving vascular and paediatric neurosurgeons, paediatric neurologists and neuro-interventional radiologists. Surgery was deemed a better option as it enabled evacuate of the haematoma and complete resection the aneurysm. Endovascular embolization was felt to be a high-risk procedure taken the location of the aneurysm. To our knowledge, however, there are no randomised controlled trials comparing the two strategies in infants of less than one-year of age, hence, the choice of the treatment of option continues to be made on a case-by-case basis.

Known post-SAH complications are reported to be extremely rare in this age group with hydrocephalus needing intervention (serial lumbar punctures or

ventriculo-peritoneal-shunts) mentioned in only 4 cases in the literature [2, 4]. Male sex and posterior circulation aneurysms were the only two recognised factors to be significantly associated with poorer outcomes [2]. In our case where the aneurysm is localised to the middle cerebral artery, swift neurological recovery was noted and complications such hydrocephalus and/or ischemic neurologic deficits were not observed. Excellent recovery in this age group has been attributed to very low incidence of hypertension and the absence of cerebral vasospasm [2, 4].

Conclusion

Infant intracranial aneurysms may differ from those in adults or even older children in location, aetiology, clinical presentation and management. In infants, non-traumatic/non-infectious aneurysms are commonly seen in the middle cerebral artery with higher risk of bleeding. Acute neurological deterioration in an infant with a spontaneous intracranial bleed should prompt aggressive evaluation with the suspicion of intracranial aneurysms as a potential cause. Early identification and immediate management should provide excellent outcomes.

Disclosure

The authors have no personal or institutional interest in any of the drugs, materials or devices described in this paper.

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